



Overview

Points To Remember About Sjögren's Syndrome

- Sjögren's syndrome is a disease that affects the glands that make moisture. It most often causes dryness in the mouth and eyes.
- The main symptoms of Sjögren's syndrome are dry eyes and dry mouth.
- Sjögren's syndrome is an autoimmune disease. In Sjögren's syndrome, your immune system attacks the glands that make tears and saliva (spit).
- Doctors diagnose Sjögren's using a medical history, physical exam, certain eye and mouth tests, and blood tests.
- Treatment differs for each person and depends on what parts of your body are affected and focuses on getting rid of symptoms.
- Living with Sjögren's syndrome can be easier by taking good care of your eyes and mouth, by protecting your voice, and understanding what medicines cause dryness.

Sjögren's syndrome is an autoimmune disease, which means that your immune system turns against the body's own cells. Normally, the immune system works to protect us from disease by destroying harmful invading organisms like viruses and bacteria. In the case of Sjögren's syndrome, disease-fighting cells attack various organs, usually in the glands that produce tears and saliva. Damage to these glands reduces both the quantity and quality of their secretions.

Sjögren's syndrome is also a rheumatic disease, which primarily affect:

- Joints.
- Tendons.
- Ligaments.
- Bones.
- Muscles.

The signs and symptoms of rheumatic diseases can include:

- Redness or heat.
- Swelling.
- Pain.
- Loss of function.

Who Gets

Sjögren's syndrome can affect people of either sex and of any age, but most cases occur in women. The average age for onset is late forties, but in rare cases, Sjögren's syndrome is diagnosed in children.

Types

Doctors classify Sjögren's syndrome as either primary or secondary. You have primary Sjögren's syndrome if you do not have other rheumatic diseases. You have secondary Sjögren's syndrome if you already have another rheumatic disease, such as rheumatoid arthritis or systemic lupus erythematosus, scleroderma, or polymyositis.

Symptoms

Sjögren's syndrome can cause many symptoms, the most common symptoms include:

- Dry eyes. If you have Sjögren's syndrome your eyes may burn or itch. Some people say it feels like they have sand in their eyes. You may have trouble with blurry vision, or bright lights, especially fluorescent lighting.
- Dry mouth. If you have Sjögren's syndrome your mouth may feel chalky or like your mouth is full of cotton. It may be difficult to swallow, speak, or taste. Because you lack the protective effects of saliva, you may develop more dental decay (cavities) and mouth infections.

Sjögren's syndrome can also affect other parts of your body, causing symptoms such as:

- Multiple sites of joint and muscle pain.
- Prolonged dry skin.
- Skin rashes on the extremities.
- Chronic dry cough.
- Vaginal dryness.
- Numbness or tingling in the extremities.

- Prolonged fatigue that interferes with daily life.

A small number of people with Sjögren's syndrome may develop lymphoma. A form of cancer, lymphoma can affect the salivary glands, lymph nodes, the gastrointestinal tract, or the lungs. If you have enlargement of a salivary gland, you should contact your doctor. Other symptoms may include the following:

- Unexplained fever.
- Night sweats.
- Constant fatigue.
- Unexplained weight loss.
- Itchy skin.
- Reddened patches on the skin.

Many of these can be symptoms of other problems, including Sjögren's syndrome itself. Nevertheless, it is important to see your doctor if you have any of these symptoms.

Causes

Researchers think Sjögren's syndrome is caused by a combination of genetic and environmental factors. Several different genes appear to be involved, but scientists are not certain exactly which ones are linked to the disease, because different genes seem to play a role in different people.

Scientists think that the trigger may be a viral or bacterial infection. The possibility that the endocrine and nervous systems play a role in the disease is also under investigation.

Diagnosis

Your doctor will diagnose Sjögren's syndrome using your:

- Medical history. Because there are many causes of dry eyes and dry mouth (including many common medications, other diseases, or previous treatment such as radiation of the head or neck), your doctor needs a thorough history.
- A physical exam. During the exam, your doctor will check for clinical signs of Sjögren's syndrome, such as indications of mouth dryness or signs of other connective tissue diseases.
- Results from clinical or laboratory tests. Depending on what your doctor finds during the history and exam, he or she may want to perform some tests or refer you to a specialist to establish the diagnosis of Sjögren's syndrome, including:
 - Blood tests to determine the presence of antibodies common in Sjögren's syndrome, including anti-SSA and anti-SSB antibodies or rheumatoid factor.
 - Other tests can identify decreases in tear and saliva production.
 - Biopsy of the saliva glands and other specialized tests can also help to confirm the

diagnosis. additional tests to see whether other parts of the body are affected.

Treatment

Treatment can vary from person to person, depending on what parts of your body are affected.

Treatments for Dry Eyes

There are many treatments you can try or your doctor can prescribe for dry eyes. Here are some that might help:

- Eye drops keep your eyes moist by replacing the natural tears. These products are available by prescription or over the counter under many brand names. Eye drops come in different thicknesses, so you may have to experiment to find the right one. Some drops contain preservatives that might irritate your eyes. Drops without preservatives usually don't bother the eyes.
- Ointments are thicker than eye drops. Because they moisturize and protect the eye for several hours, and may blur your vision, they are most effective during sleep.
- Other therapies, such as plugging or blocking the tear ducts, anti-inflammatory medication, or surgery may be needed in more severe cases.

Treatments for Dry Mouth

There are many remedies for dry mouth. You can try some of them on your own. Your doctor may prescribe others. Here are some many people find useful:

- Chewing gum and hard candy. If your salivary glands still produce some saliva, you can stimulate them to make more by chewing gum or sucking on hard candy. However, gum and candy must be sugar-free, because dry mouth makes you extremely prone to progressive dental decay (cavities).
- Taking sips of water or another sugar-free, noncarbonated drink throughout the day to wet your mouth, especially when you are eating or talking. Note that drinking large amounts of liquid throughout the day will not make your mouth any less dry and will make you urinate more often. You should only take small sips of liquid, but not too often. If you sip liquids every few minutes, it may reduce or remove the mucus coating inside your mouth, increasing the feeling of dryness.
- Using an oil or petroleum-based lip balm or lipstick can soothe dry, cracked lips. If your mouth hurts, your doctor may give you medicine in a mouth rinse, ointment, or gel to apply to the sore areas to control pain and inflammation.
- Using other therapies, such as saliva substitutes or medications that stimulate the salivary glands to produce saliva, is sometimes indicated.

Treatments for Symptoms in Other Parts of the Body

If you have extraglandular involvement, which means a problem that extends beyond the moisture-producing glands of your eyes and mouth, your doctor may treat those problems using

nonsteroidal anti-inflammatory drugs (NSAIDs) or immune-modifying drugs.

Who Treats

The symptoms of Sjögren's syndrome usually develop gradually and are similar to those of many other diseases. This means it can take time to get a diagnosis. You may see a number of doctors, any of whom could diagnose the disease and help with your treatment, including:

- A rheumatologist, a doctor who specializes in diseases of the joints, muscles, and bones.
- A primary care physician.
- An internist.
- An ophthalmologist, a doctor who specializes in caring for the eyes.
- An otolaryngologist, a doctor who specializes in caring for the ears, nose, and throat.

Usually a rheumatologist will coordinate your treatment among a number of specialists.

Living With

General Tips for Eye Care

- Don't use eye drops that irritate your eyes. If one brand or prescription bothers you, try another. Eye drops that do not contain preservatives are usually essential for long-term use.
- Practice blinking. You tend to blink less when reading or using the computer. Remember to blink 5 to 6 times a minute.
- Protect your eyes from drafts, breezes, and wind.
- Put humidifiers in the rooms where you spend the most time, including the bedroom, or install a humidifier in your heating and air conditioning unit.
- Don't smoke, and stay out of smoky rooms.
- Apply mascara only to the tips of your lashes so it doesn't get in your eyes. If you use eyeliner or eye shadow, put it only on the skin above your lashes, not on the sensitive skin under your lashes, close to your eyes. Avoid facial creams on the lower lid skin at bedtime if you are awakening with eye irritation.
- Ask your doctor whether any medications that you are taking contribute to dryness. If they do, ask how the dryness can be reduced.

Importance of Oral Hygiene

Natural saliva contains substances that rid the mouth of the bacteria that cause dental decay (cavities) and mouth infections, so good oral hygiene is extremely important when you have dry mouth. Here's what you can do to prevent cavities and infections:

- Visit a dentist regularly, at least twice a year, to have your teeth examined and cleaned.

- Rinse your mouth with water several times a day. Don't use mouthwash that contains alcohol, because alcohol is drying.
- Use toothpaste that contains fluoride to gently brush your teeth, gums, and tongue after each meal and before bedtime. Non-foaming toothpaste is less drying.
- Floss your teeth every day.
- Avoid sugar between meals. That means choosing sugar-free gum, candy, and soda. If you do eat or drink sugary foods, brush your teeth immediately afterward.
- See a dentist right away if you notice anything unusual or have continuous burning or other oral symptoms.
- Ask your dentist whether you need to take fluoride supplements, use a fluoride gel at night, or have a varnish put on your teeth to protect the enamel.

Protect Your Voice

You can develop hoarseness if their vocal cords become inflamed or become irritated from throat dryness or coughing. To prevent further strain on your vocal cords, try not to clear your throat before speaking. Clearing your throat is hard on the vocal cords. To avoid irritating your vocal cords:

- Sip water.
- Chew sugar-free gum.
- Suck on sugar-free candy.
- Make an "h" sound, hum, or laugh to gently bring the vocal cords together.

Medicines and Dryness

Certain drugs can contribute to eye and mouth dryness. If you take any of the drugs listed below, ask your doctor whether they could be causing symptoms. Don't stop taking any medicine without asking your doctor. Sometimes your doctor can make adjustments to the dose to help protect you against drying side effects or chose a drug that's least likely to cause dryness. Drugs that can cause dryness include:

- Antihistamines.
- Decongestants.
- Diuretics.
- Some antidiarrhea drugs.
- Some antipsychotic drugs.
- Tranquilizers.
- Some blood pressure medicines.
- Antidepressants.

Research Progress

Through basic research on the immune system, autoimmunity, genetics, and connective tissue diseases, researchers continue to learn more about Sjögren's syndrome. The hope is that a better understanding of the disease and its causes will lead to better treatments and perhaps

even prevention.

Some research into Sjögren's syndrome include the following:

- **Genetic studies.** Research has discovered that the *STAT4* gene variant associated with rheumatoid arthritis and lupus is also associated with Sjögren's syndrome. This suggests that variants of this gene play a crucial role in autoimmunity.
- **Immune system studies.** Researchers are investigating the various components of the immune system that may be involved in the autoimmune causes of Sjögren's syndrome. For example, studies are looking for autoantibodies that are unique to Sjögren's, and whether blood tests can be developed to detect them. Other research is investigating the role of T cells, another component of the immune system, in Sjögren's.
- **Hormonal factors.** Because Sjögren's syndrome affects mostly women, female reproductive hormones may play a role. Although studies have shown that levels of estrogen and progesterone differ little between women with Sjögren's syndrome and those without, higher levels of prolactin (a hormone that stimulates the production of milk after childbirth and the production of progesterone in the ovary) are found in women with Sjögren's syndrome. Research is also looking at how the disease affects men and women differently.
- **Prevalence of extraglandular involvement.** Studies have shown that neurological involvement and Sjögren's-related problems with the skin, including alopecia (a condition characterized by hair loss), vitiligo (a condition in which areas of the skin lose their pigment and become white), and vasculitis (a raised rash), may be more common than previously thought. Studies also indicate that identifying and treating these problems in people with Sjögren's syndrome is an important part of managing the disease. Another study shows that clinical depression is also common among Sjögren's syndrome patients, and may warrant treatment.
- **Predicting lung involvement.** Knowing who is at highest risk of certain complications can enable doctors to identify and treat these problems earlier and more appropriately. Investigators are studying proteins that are higher in people who later develop lung problems with primary Sjögren's syndrome.
- **Role of infection.** Doctors believe that infections may trigger Sjögren's syndrome in people genetically predisposed to the disease. Viral infection is under investigation as a possible trigger for Sjögren's syndrome and other autoimmune diseases.
- **Long-term relief for dry mouth.** Scientists are exploring regenerative medicine and tissue engineering strategies that may correct the underlying defects in Sjögren's syndrome.

Researchers continue to explore the genetic and environmental factors that cause Sjögren's syndrome, with the goal of creating novel diagnostic and therapeutic approaches.

The National Institute of Dental and Craniofacial Research conducts studies to help scientists understand, manage, and treat Sjögren's syndrome. If you think you might like to take part in a clinical trial, speak with your doctor or check <https://www.clinicaltrials.gov> for a listing of trials for which you may be eligible.

Related Resources

U.S. Food and Drug Administration

Toll free: 888-INFO-FDA (888-463-6332)

Website: <https://www.fda.gov>

[Drugs@FDA](https://www.accessdata.fda.gov/scripts/cder/daf) at <https://www.accessdata.fda.gov/scripts/cder/daf> [Drugs@FDA](https://www.accessdata.fda.gov/scripts/cder/daf) is a searchable catalog of FDA-approved drug products.

Centers for Disease Control and Prevention, National Center for Health Statistics

Website: <https://www.cdc.gov/nchs>

National Eye Institute

Website: <https://www.nei.nih.gov>

National Institute of Dental and Craniofacial Research (NIDCR)

Website: <https://www.nidcr.nih.gov>

NIDCR Sjögren's Syndrome Clinic

Website:

<https://www.nidcr.nih.gov/Research/NIDCRLaboratories/MolecularPhysiology/SjogrensSyndrome/>

National Institute of Neurological Disorders and Stroke

Website: <https://www.ninds.nih.gov>

American Academy of Dermatology

Website: <https://www.aad.org>

American Academy of Ophthalmology

Website: <http://www.aao.org>

American Association for Dental Research and International Association for Dental Research

Website: <http://www.iadr.org>

American College of Rheumatology

Website: <https://www.rheumatology.org>

American Dental Association

Website: <https://www.ada.org>

American Autoimmune Related Diseases Association, Inc.

Website: <https://www.aarda.org>

Arthritis Foundation

Website: <https://www.arthritis.org>

Lupus Foundation of America

Website: <https://www.lupus.org>

The Myositis Association

Website: <https://www.myositis.org>

National Organization for Rare Disorders

Website: <https://www.rarediseases.org>

Scleroderma Foundation

Website: <https://www.scleroderma.org>

Scleroderma Research Foundation

Website: <https://www.srfcure.org>

Sjögren's Syndrome Foundation, Inc.

Website: <https://www.sjogrens.org>

If you need more information about available resources in your language or other languages, please visit our webpages below or contact the NIAMS Information Clearinghouse at NIAMSInfo@mail.nih.gov.

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